CASE REPORT

A New Clinical Entity Mimicking Meningioma Diagnosed Pathologically as Rosai-Dorfman Disease

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Abstract—Sinus histiocytosis with massive lymphadenopathy (SHML, Rosai-Dorfman disease), is a well-defined histioproliferative disorder. The extranodal type of the disease is not uncommon, but CNS involvement is extraordinary. Moreover, the isolated extranodal intracranial manifestation of the disease is extremely rare. Only a few cases of the latter form have been reported. We present a case of meningioma-like intracranial lesion, which after total resection, pathologically and immunohistochemically proved to be an isolated form of the Rosai-Dorfman disease. No concurrent nodal involvement was found, even after a detailed postoperative diagnostic work-up.

Two years follow-up disclosed full recovery from the CNS symptoms, while no other SHML signs were noted. The extranodal form of the Rosai-Dorfman Disease, nowadays has been enlisted in the differential diagnosis of the space occupying intracranial and skull base lesions. However, a number of questions, dealing with the preoperative diagnosis, the surgical strategies, as well as the natural history, the prognosis of the disease, and the possible need for adjunctive therapy, remain obscured. Additional reports are useful to better understand this rare type of disease and may lead to the establishment of a more profitable management.

In 1969, Rosai and Dorfman described a benign histioproliferative disease which is currently classified as an idiopathic histiocytosis. They named the entity sinus histiocytosis with massive lymphadenopathy (SHML or Rosai-Dorfman disease).¹ SHML is characterized clinically by massive cervical lymphadenopathy, fever, leukocytosis with an increased erythrocyte sedimentation rate¹ and pathologically by enlarged lymph node sinuses containing large histiocytes whose cytoplasm contains well preserved lymphocytes, a phenomenon referred to as lymphocytophagocytosis or emperipolesis.²

More than 400 cases of SHML have been reported since its original description. This disease can also be found in other lymph nodes or extranodal organs.² The extranodal type of the disease is not uncommon, but CNS involvement is extraordinary. Moreover, the isolated extranodal intracranial manifestation is extremely

rare. To our knowledge, five cases of isolated extranodal intracranial involvement have been reported.^{1,3-6}

We describe a disease confined to a dural-based intracranial lesions mimicking parasagittal meningioma which pathologically was diagnosed as sinus histiocytosis with massive lymphadenopathy, while a 2-year follow-up confirmed the isolated, extranodal, intracranial type of the disease. Thus we believe that clinically it must be a new entity.

CASE REPORT

A 62-year-old Greek man was admitted to our clinic suffering from tonicoclonic seizure for first time in his life. He had been previously healthy without a significant personal or family history.

Physical Examination

The patient was alert and cooperative and complaining of headache. Neurological examination revealed expressive dysphasia and right hemiparesis, predominantly in the arm. No other neurological deficit was found. He was afebrile with no lymphadenopathy.

In routine investigations hematological and biochemical studies were normal, the skull x-ray was normal but a diaphragmatocele was found on the chest x-ray. Thus, we decided to continue the investigations with thorax and abdomen computer tomography which showed that the mediastinum and the retroperitoneal space were free from lymphadenopathy (Figs. 1 and 2). The liver, spleen, kidneys, and pancreas appeared to be normal. The diagnostic work-up was completed with Cranial computerized tomography with and without

contrast media administration, and brain magnetic resonance imaging scan. The CT scan disclosed a well circumscribed enhancing mass lesion in the left parietal lobe with a base of attachment along the falx. Marked edema was also noted (Fig. 3). The appearance of the lesion was better defined to be dural-based by the MRI scan, having the same radiological characteristics with the meningiomas (Figs. 4–7).

The patient was diagnosed preoperatively as having parasagittal meningioma and was put on Dexamethazone.

Figure 1. Lymphadenopathy-free mediastinum. Thorax CT scan.

Figure 2. Retroperitoneal space free from lymphadenopathy. Abdominal CT scan.

Figure 3. Contrast-enhanced CT scan. A well circumscribed enhancing mass in contact with falx. Note the extended parietal lobe edema.

Figure 4. Contrast-enhanced T₁-weighted MRI (axial view) confirming the enhanced lesion, and the extension of the tumor to opposite parietal lobe.

Figure 5. The mass lesion being isointense to the brain. T₁-weighted MRI, axial view.

Figure 7. Axial T₂-weighted MRI, confirming the isointensity of the mass lesion to the brain tissue.

Figure 6. The mass in contact with falx and superior sagittal sinus (coronal, contrast-enhanced, T₁-weighted MRI).

Operation

The mass was exposed via a left parietal craniotomy. The tumor appeared to be <<en plaque>> meningioma (a meningioma that has a pancake-like appearance), xanthochromic, firm, well circumscribed, and adherent to the cerebral falx and to the wall of the superior sagittal sinus

but any obvious matrix was not detected. The tumor was radically excised in very small fragments.

Pathological Examination

Microscopic examination showed a mixed cellular population composed predominantly by polymorphous aggregates of large histiocytes, lymphocytes and some plasma cells. One of the most important findings was the phenomenon of empiropolesis or lymphocytophagocytosis (Figs. 8 and 9). Also the S-100 staining in the histiocytic elements was noted.

Postoperative Course

After surgery patient had an uneventful course. His headaches and the right hemiparesis ceased, while the dysphasia persisted. He was discharged on the ninth postoperative day without any medication.

Follow-Up

One month postoperatively the patient had completely recovered from his expressive dysphasia. One year after the operation the patient was found to be in good health without findings from the brain CT and MRI scan. Twenty-four months after surgery the patient was found to be in excellent condition, without neurological or any other systems findings, and free from any

Figure 8. Photomicrograph (magnification $\times 40$) of the mass lesion. Section showing fibroinflammatory tissue.

medication. Brain CT scan (Figs. 10 and 11) and MRI (Figs. 12–14) revealed no evidence of recurrence. CT of the thorax and the abdomen ruled out any lymphadenopathy (Figs. 15 and 16).

DISCUSSION

Since its initial description by Rosai and Dorfman, sinus histiocytosis with massive lymphadenopathy (SHML) has become an established clinicopathological entity.⁵ Our experience being in agreement with other authors,^{3,7,8} has led us to believe that the CNS lesions of Rosai-Dorfman disease may mimic meningiomas in clinical and radiological appearance as well as in surgical findings.

Evaluating the results from the follow-up of our case, where typical clinical and laboratory features of SHML are entirely absent, we sustain our opinion that the term SHML is inappropriate for cases of isolated extranodal intracranial manifestation in case of not detecting other lesion in the follow-up. The intracranial extranodal disease is not a common feature of SHML as previously believed.²

The involvement of the disease does not respect the anatomical structures of the skull base^{4-7,9} and the routine preoperative diagnostic work-up fails to reveal the precise nature of the tumor. Treatment with radiosurgery of what was diagnosed radiologically as a skull base meningioma might be a mistake.¹⁰ Until now only a biopsy correlates the intracranial lesion with the Rosai-Dorfman disease and suggests the denotation for

Figure 9. Photomicrograph (magnification $\times 100$) of the mass lesion. Section showing foamy histiocytes with emperipolesis of lymphocytes and plasma cells.

Figure 10. Postoperative CT scan showing the craniotomy and the underlying scar tissue (24 months postoperatively).

Figure 12. T₁-weighted MRI, axial view. The existence of postoperative scar tissue in the operated area and the perifocal lucidness (2 years postoperatively).

Figure 11. Postoperative CT scan with evidence of no recurrence (2 years postoperatively).

Figure 13. The scar tissue and the gliosis of the surrounding brain parenchyma is shown in this porton density MRI.

further investigations to detect other manifestations of the disease.

The surgical excision, radical if possible, must be the appropriate treatment. Finally it seems to be necessary to get more familiar with this disease and we need more long-term and broad spectrum follow-up investigations, to detect a possible local recurrence, and to be as certain as possible if complementary treatment (radiotherapy, chemotherapy) would be beneficial in these cases.

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Figure 14. T₁-weighted magnetic resonance image, coronal view, confirming the absence of any recurrence (2 years postoperatively).

Figure 15. Postoperative CT scan (24 months), mediastinum free from lymphadenopathy.

Figure 16. Abdomen postoperative CT scan (24 months). The retroperitoneal space free from lymphadenopathy.

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